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THE EFFECTS OF CONGENITAL
RUBELLA.

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THE EFFECTS OF CONGENITAL RUBELLA

Children receiving services in programs for the deaf-blind are not simply visually and hearing impaired. Additional complications beyond deaf-blindness can result from fetal infection, faulty genetic control over embryological development and postnatally acquired diseases. However, since the Deaf-Blind Regional Centers were established in the United States largely as a consequence of the 1964-65 epidemic of rubella which left 30,000 children in this country with congenital malformations, I would like to share with you some findings regarding the clinical manifestations of this disease.

For many years deafness, cataracts and congenital heart disease were considered the congenital rubella syndrome. However, recent research indicates that the consequences of fetal infection are much more diverse. During the infection period, virtually every tissue in the fetus, including that of the eye, inner ear, heart and brain, is in danger. Possible impairments range from sensorineural deafness (bilateral or unilateral, mild to profound), congenital heart disease, cataracts, glaucoma, high myopia, rubella meningoencephalitis, with resultant degrees of psycho-motor retardation with intellectual or motor impairment, various behavior abnormalities (often autistic in quality), central language impairment or typical spastic cerebral palsy.

Another pre-1964 misconception is that congenital rubella is a condition of static, structural abnormalities. It is now known that rubella is a continuing infectious disease. Children with congenital rubella may excrete virus from birth to 18 months of age. Futhermore, problems attributable to rubella infection in utero may not become apparent until the adolescent years. This includes endocrine and neurological disorders and possibly other diseases of late onset, such as malignancies.

There have been occasional anecdotal reports of children with congenital rubella who have developed progressive hearing loss as they reached later childhood and adolescence. The reasons for this are unclear but fortunately it rarely happens. There is no indication of progressive vision loss but it should be noted that the eyes of both normal children and those with congenital rubella undergo a growth spurt in globe diameter during adolescence. Since myopia can result, careful vision testing should be performed on at least an annual basis.

The Rubella Project in New York City currently has approximately 600 children in active longitudinal follow-up. Within this population 6 children with overt diabetes mellitus have been identified. However, an Australian study of older persons with congenital rubella found the incidence of diabetes mellitus to be 20%. Statistically, then, there

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seems to be an increased probability of the child with congenital rubella developing an endocrine disorder.

In general, the earlier in pregnancy that rubella is contracted, the greater the likelihood that brain injury will occur and that it will be severe. Futhermore, researchers at the Rubella Project have pointed out the continuously evolving character of the neurologic aspects of children with congenital rubella. Still, the plasticity of the central nervous system and its capacity for utilization of relatively undamaged portions of the brain should not be underestimated.

In a study of 74 children with mental retardation secondary to congenital rubella 21 considered retarded at 4 to 5 years of age were no longer classified as retarded at 8 to 9. Children seen during infancy with extreme irritability, tremulousness and opisthotonic behavior reflect an ongoing meningoencephalitis secondary to rubella infection. With the clearing of this encephalopathic process, which may take place between 3 and 5 years of age, there is often a gratifying spurt in development and especially in the areas of ambulation and self-care skills. Language development lags behind because of the frequent association of severe or profound hearing loss and "central language impairment" with mental retardation.

There are a number of disturbing facts about the neurological picture in early adolescence which need to be discussed. First, seizure disorders have been appearing in this population with greater frequency and in a much higher percentage than would be expected in a random population. Second, pediatricians should be alert for the appearance around the time of puberty of progressive rubella panencephalitis. No child followed by the Rubella Project has developed this problem, but it has been observed in a group of children in California with congenital rubella. The disease is characterized by progressive ataxia, behavioral and intellectual deterioration and seizures, leading to death in two of eight patients.

Rubella children may not develop behavioral pathology, but there is an increased risk of their developing some type of behavioral disturbance. In a study of 243 children aged 2 1/2 - 4 enrolled with the Rubella Birth Defect Evaluation Project in New York City, half of the children showed no behavioral pathology. All the remaining children fit into one or more psychiatric categories, including cerebral dysfunction, childhood psychosis, schizophrenic adjustment, reactive behavior disorders and mental retardation. Other diagnostic categories may be added at later ages when personalities become capable of complex reactive patterns.

The diagnosis of cerebral dysfunction is made only when there is

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independent evidence of neurological disease as well as a coexistent state of behavior disorder characterized by symptoms of brain damage, such as hypo or hyperactivity, attention abnormalities and impulse disorders. Eight children or 3.3% of the study population were found to have this disorder but it is possible that more will develop behavioral aberrations and limitations later in life when the children are faced with increased demands for impulse control and flexibility of functioning.

Reactive behavior disorders develop as a consequence of stress. The assumption is that with a beneficial change in the external circumstances and environmental handling there will be a corresponding improvement in the child's behavior. An example might be difficulty at home but not in school. 37 children or 15% of the total sample had reactive behavior disorders. The authors of the study caution that while the incidence does not seem particularly high, there is reason to fear that excessive stress may be imposed on handicapped youngsters as they grow older.

Psychosis is diagnosed when the child's behavior is disorganized and out of touch with reality. Schizophrenic adjustment implies the presence of deviations in thought processes, affect, interpersonal relatedness, motility and speech patterns which fall a little short of a psychotic state. None of the rubella children in the study gave evidence of a psychosis or childhood schizophrenia other than autism. The primary criteria for this diagnosis are indications that the child has an affective defect and tries frantically to maintain sameness in his environment. Possible symptoms reported by Kanner (1943) include inability to relate in the ordinary way to people and situations from the beginning of life; (2) monotonously repetitive verbal utterances which do not convey meaning to others; (3) excellent rote memory but inability to use language in any other way; (4) viewing outside events (food, noise and motion) as an intrusion; (5) limited spontaneous activity; and (6) good relation to objects with which the child can play happily for hours. The behavior of ten youngsters corresponded with Kanner's criteria with the additional finding of mental retardation in all but one of them. Another eight children exhibited a significant number of autistic behaviors and were diagnosed as exhibiting a "partial syndrome of autism." We can see just how high the prevalence of autism is in this sample of rubella children when we compare figures with a 1970 Wisconsin study in which infantile autism was found in .7 per 10,000 youngsters and childhood schizophrenia, including autism, in 3.1 per 10,000. In contrast, 10 out of 243 yields a prevalence rate of 412 per 10,000 for the core syndrome of autism and 329 per 10,000 for the partial syndrome, yielding a combined figure of 741 per 10,000.

The term mental retardation refers to "subnormal general intellectual

functioning which originates during the developmental period and is associated with impairment of either learning and social adjustment or maturation or both." 91 youngsters or 37% of the total sample were retarded as compared with 2-3% of the general population. In addition, the severely retarded comprised the largest group and the combination of those who were severely or profoundly retarded represented almost half of the total group of retardates (43 of 91). This is in sharp contrast to the fact that the vast majority of retarded youngsters falls into the borderline or mild group.

It should be noted that many of the rubella children had more than one psychiatric diagnosis. Of the 125 children with a psychiatric problem, 99 (79.2%) had a single disorder which for the most part was a reactive behavior disorder (30 children) or some degree of mental retardation (65 children). 26 youngsters were given multiple psychiatric diagnoses. Five had reactive behavior disorders and mental retardation. Nine were mentally retarded and autistic and six others combined mental retardation and a partial syndrome of autism. The remaining six all had cerebral behavior disorders and one also showed a partial syndrome of autism.

It is also illuminating to study the relationship between physical impairment and psychiatric status. Of the 118 children with no psychiatric disorders, 40 had no physical defects at all and 41 had only one area of physical defect. As the number of areas in which defects were present increases, the number of children without psychiatric disorders decreases and a youngster becomes more and more likely to display coexistent behavioral pathology. Still, the fact that 37 children had two or more areas of physical defect and were free of psychiatric disorders cautions against assuming a necessary relationship between physical impairment and psychiatric status.

The authors also studied the relationship of kind and number of physical defects to psychiatric diagnosis. Hearing defects were most common in this group of youngsters. In fact, the most important consequence of rubella is deafness. In the same 243 rubella children studied 65 (26.7%) had hearing defects alone and a further 112 youngsters had a hearing loss in combination with other defects. Thus, almost 3/4 of the children presented deafness as a major disability. Still, only 41.7% of those with hearing loss alone or in combination with either or both cardiac and neurological defects had psychiatric disorders. There were far fewer cases of visual handicap alone (4) or in combination with either or both cardiac and neurological defects (59), but 81.8% of children with these handicaps exhibited behavioral pathology. Significantly, 76.2% of children with only hearing and visual defects (14) or combined with one other physical handicap (28) had psychiatric disorders.

This makes the "deaf-blind" rubella child particularly vulnerable.

The authors also wanted to know if there was such a thing as a "behavioral rubella syndrome." Therefore, in considering areas of behavioral disturbance, the presence or absence of psychiatric pathology was not taken into account. The focus was rather on specific deviations beyond the range of normalcy.

46% of the rubella children had sleep problems, an unusually high frequency even for preschoolers. Many children went to bed very late, woke up during the night, walked around the house at night after being put to bed or went into their parents' bed. Interestingly, nightmares don't seem to be a problem in this population as the mothers rarely reported that their children woke up screaming and crying.

53% of the rubella children had difficulties in the area of feeding and eating. The children did not progress smoothly from drinking fluids to eating solids and they often refused to learn to eat. A possible physical basis for these problems is difficulty in swallowing, a common neurological sign found in the group as a whole.

More than 1/4 of the children exhibited problems in dressing. Some of these difficulties seem to be due to the fact that they found it difficult to acquire self-care skills. Motor incoordination is also a factor to consider. However, other youngsters insisted that parental servicing be perpetuated. Others had phobic reactions to clothing and seemed especially distressed by tight garments being pulled over their heads and by having to keep their feet still while shoes were being put on and tied. These problems could be due to an excessively high activity level and an unwillingness to remain still. Other children may have developed a very low threshold to tactile sensation to compensate for visual and hearing deficits. Such hyperactivity may also be the first manifestation of cerebral dysfunction.

37% of the population had difficulties with elimination. This was attributed to retardation, irregularity in biological functioning and problems in oral communication.

Mood difficulties were found in almost 3/4 (72%) of the children. Separation anxiety and clinging or withdrawn behavior were especially prominent. Mothers reported temper tantrums, screaming without provocation and affective deviances or mood disturbances at home. Possible causes include cerebral dysfunction or a high degree of frustration felt by children with hearing deficits who can't express their needs and desires in speech. Moreover, their hearing problems allow them to receive only incomplete signals from their parents and their partial

understanding of parental demands makes it that much harder to distinguish between behavior that will be rewarded and that which will be reprimanded.

Discipline was one of the greatest problems with 64% of the children considered deviant in this area. Sometimes parents were reluctant to punish a child for infractions of rules of which he might not even be aware. At other times they could not bring themselves to punish a child who was handicapped.

Often it was the consequences of disruptive and destructive behavior that was worrisome rather than the fact that the behaviors were undisciplined. If behavior did not threaten a child's personal safety or somebody's valuable personal property, it could be ignored. Otherwise, parental vigilance and control was mandatory. This was not overprotection but simple realism. Still, a child who has had severe motor restrictions placed on him might come to experience life as a series of prohibitions and he might, therefore, acquire a number of defensive behaviors such as negativeness or fearfulness or even a sense that his parents will always take care of him.

55% of the children displayed deviant motor activity. Interestingly, parental complaints in this area usually centered on examples of hyperactivity. In contrast, hypoactivity was seen as an example of good behavior. The child who sat playing quietly by himself for hours was the good child. Parents did not see this behavior spontaneously as a sign of unusually little activity. Other behaviors such as peculiarities of gait, motoric incompetence and drooling were not seen as especially problematic by the parents. This may be because the parents did not see these problems as warranting primary attention. They may also have developed lowered expectations based on their recognition of their child's limitations in motor competence.

2/3 of the children exhibited habits and rituals. This may be related both to the high incidence of children with sensory defects and the high proportion of mentally retarded children in this study. Rubella youngsters exhibited "blindisms' such as poking their eyes, waving fingers before their eyes, rocking their head sideways and staring at the light. Other rituals in the rubella population resembled those found in mentally retarded children. Among these behaviors are arm flapping, rocking and other repetitive aimless activities. The rubella children also tended to smell objects such as toys, pages of a book and people's feet. These were unacceptable behaviors to the parents, especially since they quickly identified a child as deviant.

Somatic reactions were relatively few and consisted merely of gastrointestinal reactions such as gagging and vomiting in response to unpleasant smells. Sometimes children with intact sensory modalities exhibit a hyperactivity to smell. In the rubella children with faulty

vision and hearing, it may be that the sense of smell may be more highly developed. This would cause them to gag more frequently in response to unpleasant odors.

83% of the rubella children had speech difficulties. The relation between hearing loss and speech disturbance is, of course, logical. Still, children without defined hearing deficits also had speech defects. These deviations may be transitory maturational lags or they may be the earliest indications of cerebral dysfunction.

More than 50% of the children showed difficulty in social relationships. These problems included aggressive acts such as hitting, kicking, biting and withdrawal from other children. Parents were more likely to be concerned about their child's interpersonal behavior than about other aspects of his functioning. A normal child of the same developmental age can safely fight with a peer but it is a matter of some concern if a larger, stonger child acts agressively toward younger children. Withdrawal from others did not cause much concern among parents of preschoolers.

Problems in formal or academic learning situations were not widely reported by parents of preschool rubella children since few formal learning demands were imposed on them and they had not yet reached the age at which academic demands would be made. Still, there is much evidence that learning problems show up as the children become older.

Other behavioral deviations such as behaviors reflecting immaturity, fear and other isolated symptoms were not widely reported by parents. This probably happened because parents adjusted their expectations to a lower level and could no longer judge age-appropriateness. Almost half of the parents reported that their children had specific fears such as of water, thunder, cars and masks. What was usual was their high incidence and the intensity with which they were expressed. At the same time parents reported that their children lacked an ordinary sense of caution and did not exercise proper fearfulness in regard to experiences that had already proved harmful. One youngster repeatedly jumped from the top of a flight of stairs, expecting his parents to catch him each time.

Autistic behavior is one of the possibilities that the person dealing with rubella children must be aware of. Kanner described children who from birth gave little evidence of ability to relate to people and who displayed an obsessive desire to maintain sameness. Their capacity for spontaneous activity was limited, although they could play happily with objects for hours. They also failed to look directly at people and did not make visual contact with the eyes of the person attempting to attract their attention. Interestingly, none of these children had sensory defects, their intelligence levels were normal and there were no significant neurological findings.

Maternal attitudes toward autistic rubella children were compared with those of mothers with nonautistic rubella children. The results were that there was no difference between the two groups in the perception or denial of the physical consequences of maternal rubella and no difference in maternal attitudes. There was no evidence of "refrigerator parents," schizophrenogenic mothers or greater vulnerability to autism in some socioeconomic groups.

The researchers also felt that the striking individual differences among the retarded contradicts the speculation that autism is a variety of retardation. Any sensory defect can not explain retardation. Since all the children were at risk for prenatal invasion of the CNS by rubella virus, it seems reasonable to speculate that the autistic children were brain-damaged. Other studies showing CNS dysfunction in 8 of 33 rubella children and a high percent of encephalopathy (84%) and organic brain damage (56%) in 25 autistic, retarded children lend support to this view.

The researchers were very interested in the kind of relationship that existed between the child and his or her parents. This is often poor since a handicapped child's lack of emotional expressiveness or poor developmental progress causes the parents to feel that they are not receiving enough from the child to warrant giving him much attention. Yet the parents are expected to give him or her so much more than a normal child might require.

The parents had a clear understanding of their youngster's sensory defects and physical handicaps but were very confused about behavioral consequences. For example, some did not immediately understand that a hearing loss affects the ability to speak and other parents who knew their children were brain damaged did not know that problems such as hyperactivity, aimless behavior,attentional difficulties and impulse disorders could be expected to arise.

This lack of understanding may be due to the fact that they received information from a large variety of sources. Parents of multihandicapped children carry out many pilgrimages to different clinics and doctors. There they often receive advice which seems to be--and often in fact is--contradictory. Each specialist, addressing himself to one aspect of the child's problems, may recommend procedures that do not take another need of the child into consideration. The parents may also be confused by information received at various stages of the child's development. If, for example, all they were told at the original examination was that their child had some visual difficulties, they may not be able to assimilate the fact that rubella virus is also responsible for speech defects.

A number of parents simply ignored certain defects because the multiplicity of handicaps was overwhelming. Some parents claimed that their child was simply profoundly deaf; others said their child had a hearing or vision defect and gave a quantitative appraisal of the defect but didn't really understand what this meant in terms of daily living.

On the whole, the parents were most concerned about profound deafness with consequent lack of speech. They felt that without speech the child would never be able to live independently. This concern grew as the child became older.

39% of the 243 preschool rubella children were first-born. Thus, in many cases, the birth of a damaged child came at a point when insecurities about personal competence often came to the fore. Some were so shaken that they resolved not to have any more children Another disturbing discovery for these inexperienced parents was that they had neither time nor the ability to develop skills in their babies. Many parents exhausted themselves emotionally trying to deal with that aspect of their child's defect that hit them the hardest, usually the lack of emotional expressiveness or deafness. In large families with older children, the stress of having a rubella child was lessened by the availability of siblings to assume responsibility during the mother's absence. Also,the fact that other children were not damaged mitigated disappointment and worries over the defective child.

Parents were torn over the issue of how much time should be given to their rubella child in comparison with normal siblings. Some parents felt that if the child couldn't learn, there was no point in insisting that he or she make an effort. Others felt they must spend all their time with the child helping him to master what was necessary.

Emotional problems also developed between husband and wife. Fathers who generally spent less time with their very young children than mothers became concerned that their son would not develop a proper identification as a male and accused their wives of contributing to the problem. Fathers consequently pushed hard for certain roles and play activities for their sons even though they may not have felt it necessary to do this with a normal child.

It was also difficult for the parents to learn of additional handicaps each time the child was re-examined. This was due to the ongoing nature of the rubella infection as well as the fact that some abnormalities, such as hearing loss, could not be accurately evaluated in a very young child.

Another source of concern and embarrassment was the fact that the

children showed little or no progress in overcoming handicaps despite the many efforts of the parents.

Some parents felt guilty,in varying degrees, about having a handicapped child; others were able to take some comfort in referring to a specific disease and a highly publicized epidemic. In some cases, the disruption in the home environment was counterbalanced by the fact that parents used their energies productively in finding and securing services for their child.

The parents had their hands full coping with discipline problems. Their rubella children were frequently quite stubborn and needed to be physically restrained from what they wanted to do. This method becomes more difficult to use as the language-deficient child grows older. Many parents complained about the destructive consequences of hyperactivity. Children seemed at times to be in perpetual motion while breaking objects, turning over furniture and throwing things on the floor. The researchers felt this was an unmotivated consequence of the child's activity level.

There were many disagreements between the mother and father or inconsistencies in the same parent regarding how much to expect from the child. In general, the greater the number of defects, the more permissive the parents became. Parents were sometimes suprised to learn that their child not only accepted discipline at school, but also became more responsive in learning situations after discipline was accepted.

Two-thirds of the children had unusual habits and rituals such as mouthing and chewing objects, spinning in circles, plucking eyelids, gazing at lights, smelling everything and head-banging. In fact, parents didn't spontaneously mention these behaviors to the interviewers since they couldn't imagine they might be part of the rubella syndrome. Instead, they blamed themselves, believing that their handling techniques caused the strange behavior. The fact is, however, that the stereotyped and ritualistic behavior in these children reflect mannerisms typical of children with visual and hearing defects or may be consequences of retardation and brain dysfunction.

The parents hoped that eventually their rubella children would be able to be involved in the normal world, even if on a limited basis. Hopefully, future developments will not make realization of this goal unlikely. Still, a whole new set of stresses will arise when the preschoolers reach the age of compulsory school attendance. The other children may reject him or her and the child may respond with withdrawal, aggression and other defensive maneuvers. Then

there is the problem of educational placement and finding a facility that provides the best learning environment but that is not so "special" that s/he fails to develop skills for dealing with the normal world.

The parents did not expect the clinic or remedial services to be concerned with their family problems. Nor did they view the hospital clinic as a place to seek guidance for coping with the rubella child's behavior problems. In sum, then, the analysis of the services given to rubella families indicates, among other things, that a great need exists for coordinating the health services a child receives and for centralizing information given to parents.

*The fact that there was an overrepresentation in the study of children from Puerto Rican families (17.7% in a city where Puerto Ricans comprise 7.9% of the population) informs us that social factors play an important role in congenital rubella and its sequelae. Overcrowded living conditions increased the likelihood that pregnant Puerto Rican women were at greater risk of contracting rubella during the 1964 epidemic. In addition, there was no special effort made to alert these Spanish-speaking women of the danger they were in until it was too late for an abortion. Of course, at that time abortion was illegal if there was no threat to the mother's life and it required more sophistication and money to obtain an abortion than many low-income families could command.

It may also be helpful to know a few other statistics about the parents. 62.8% were White (non-Puerto Rican); 15% were Negro. 56.4% were Catholics; 16.9% were Protestant and 12.1% were Jewish. The vast majority of the mothers were housewives and the fathers' occupations ranged from unskilled and semi-skilled to professionals.

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